A CASE OF SPRENGEL'S DEFORMITY.[1]

By George Fetterolf, A.B., M.D., Sc.D.,
ASSISTANT PROFESSOR OF ANATOMY, UNIVERSITY OF PENNSYLVANIA,

AND

John Hancock Arnett, A.B.,
CLASS OF 1916, MEDICAL DEPARTMENT, UNIVERSITY OF PENNSYLVANIA.

(From the Laboratory of Anatomy of the University of Pennsylvania.)

This article was written to put on record a case in which a complete autopsy was obtainable. For discussion of the etiology, symptomatology, etc., of the condition the reader is referred to Horwitz's article, mentioned below.

Definition. Congenital elevation of the scapula is an affection characterized by an upward displacement of one or both scapulæ, with rotation upon the sagittal or frontal axis. Alterations in shape, size, and density of the bone exist.

Historical. This condition, both on account of its rarity and because of its puzzling etiology, has been a source of interest for fifty years, the first case recorded being that of Eulenberg's in 1863. In 1891 Sprengel described four cases and advanced a plausible theory to explain the disease; since then the condition has been called "Sprengel's deformity" or "congenital elevation of the scapula." In view of the fact that the high position is not due to elevation but to failure of descent, possibly better terms would be "undescended scapula," "congenital high position," or "scapula alta congenitalis." An analogous instance in terminology is the undescended testicle, which we do not speak of as "congenitally elevated."

Frequency. Up to 1904, 100 cases had been reported. In 1899 Honsell reported the first case of bilateral disease, and up to 1908 14 such cases had been described in the literature. Horwitz,[2] in 1908, reported 16 cases himself and collected 120 from the literature, bringing the total up to 136. Less than 20 have been characterized by the presence of an associated bony arch, as found in the case we are reporting.

Physical Examination. The condition was found by us to exist in a body in the anatomical laboratory of the University of Pennsylvania. The subject was a male negro, about forty years of age, of medium size and good muscular and general development. A notable exception to this general statement was found in the right arm, which was about 3 cm. less in circumference than the left at both the arm and the forearm. In addition to being less developed it could be raised but slightly above the horizontal, this restriction being due to a limitation in movement of the scapula. Surface examination showed that the right scapula was about 6 cm. higher than the left, and between its vertebral border and the spines of the vertebræ a bony lump could be felt.

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[1] Read before the Section on General Medicine of the College of Physicians of Philadelphia, March 23, 1914.

[2] Amer. Jour. Orthop. Surg., 1908–09. vi, 260.

Bony Anatomy. The scapula differed from most of the cases hitherto reported in that, with the exception of the scapular index, it was approximately normal in size and showed pronounced roughenings for the attachment of strong muscles. On the vertebral border just below the spine was a triangular facet (Fig. 2), which faced forward and inward and articulated with a bony projection from the sixth cervical vertebra. As is common in Sprengel's disease, the bone had a scroll-like forward curve superiorly (Fig. 3), causing the upper border to project forward as a knife-like edge. The spine made less than a right angle with the vertebræ below it, so that it pointed out and down from the midline of the body instead of out and up, as is usually the case; this latter condition is found normally in the fetus.

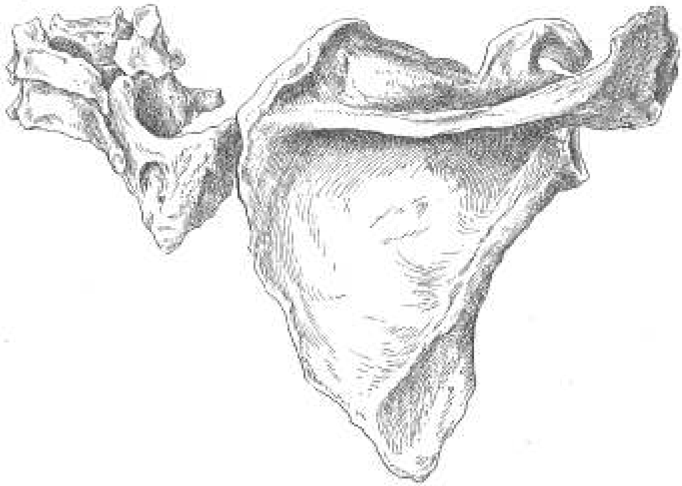

Fig. 1.—Dorsal view of scapula and fifth and sixth cervical vertebræ, showing articulation, exostosis, and spina bifida.

The Vertebræ. The scapula articulated with an exostosis from the sixth cervical vertebra, this growth causing the lump felt on external examination, and the articulation being partly responsible for the restricted upward movement of the arm.

This osseous growth consisted of a strong, markedly curved process of bone (Fig. 1, 2, 3), about 4 cm. in length, which arose from the right lamina of the sixth cervical vertebra. Viewed from the left it was seen to be roughly triangular, the apex being attached to the lamina and the base being free and directed well toward the right. Both the pedicle and lamina of the right side of this vertebra were much thickened. The growth measured about 2 cm. at the apex and about 5 cm. at the base. Its left side was separated from the vertebral spine by a narrow fissure (Figs. 1, 3), which extended clear to the spinal canal, the latter thus being open posteriorly, while the right side presented a deeply concave groove (Figs, 2, 3), for the accommodation of the semispinalis and rotatores and multifidus spinæ muscles. The upper half of the base had a distinct articular surface, which formed a true joint with the facet upon the mesial border of the scapula. This articulation was of the partially movable type, and was furnished with a capsule, the opening of which allowed the escape of fluid.

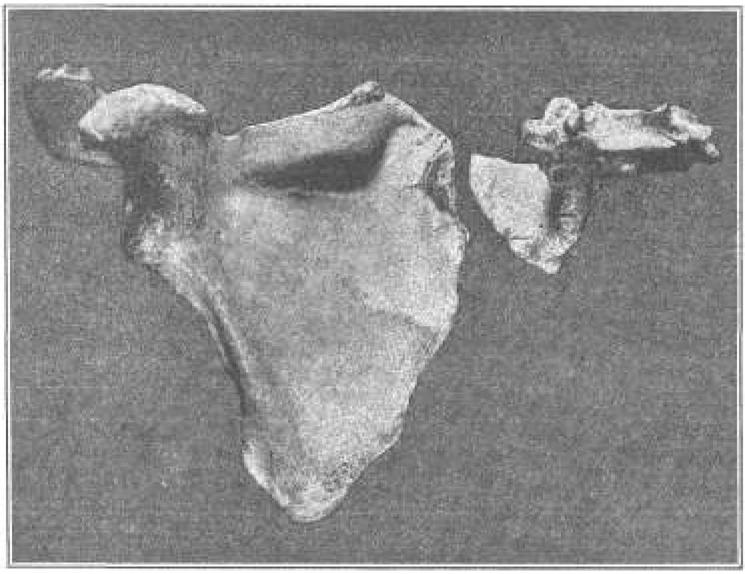

Fig. 2.—Ventral view of scapula and sixth cervical vertebra, showing the articular facet on the former and the exostosis on the latter.

A noteworthy point in regard to the vertebræ was that the right laminæ of the fifth and seventh, as well as the sixth, cervical vertebræ had not joined those of the left side to complete the arch; a condition of spina bifida (Fig. 1) was present in these three bones. The right lamina of particularly the fifth cervical vertebra was poorly developed, there being a gap (Fig. 1) of $\frac{3}{4}$ cm. posteriorly, which in the articulated state was filled in to some extent by that greatly hypertrophied right lamina of the sixth, from which sprang the osseous growth already described.

Muscular Anatomy. These anomalies of the skeleton were associated with striking deviation from the normal in the adjacent musculature (Fig. 4), the muscles involved being the trapezius, the levator anguli scapulæ, the rhomboideus major and minor, as well as several anomalous slips.

The trapezius: This muscle was represented by two portions, an upper and a lower, the former being large and very muscular and the latter small, atrophic, and composed mainly of fibrous tissue (Fig. 1). The former was so well developed that it gave the man a square, bull-necked appearance on the right side.

The upper part had the usual origin from the occiput and the cervical spine, while the lower arose from the upper thoracic vertebræ. At the first thoracic vertebra the upper muscular portion ended abruptly, and at the point where it crossed the superior angle of the scapula it became densely aponeurotic; here it overlay a bursa. The insertion was normal, with the exceptions that it extended farther medially on the clavicle than usual, and that the insertion into the acromion was thicker and broader than normal, as well as unusually aponeurotic.

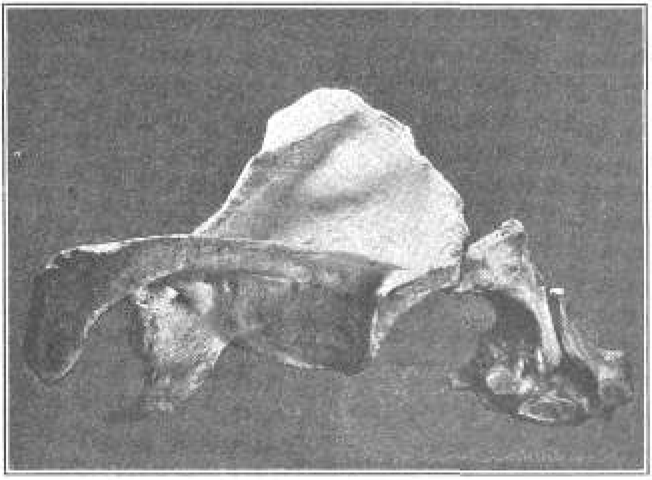

Fig. 3.—View from above of scapula and sixth cervical vertebra.

The inferior and smaller slip lay below the level of the spine of the scapula. It arose aponeurotically from the vertebral border of the scapula, the vertebral spines and the aponeurosis lying between by means of tendinous fibers, which were interwoven with superficial fibers representing the rhomboids. Under the aponeurotic origin a muscular origin appeared, the fibers intermingling with fibers of the rhomboids, and having a common origin with them from the seventh cervical and first thoracic vertebræ. It inserted into both lips of the spine of the scapula to within one inch of the acromial tip, thus encroaching upon the origin of the deltoid, and causing that muscle to attach to the spine below the lower lip.

Subtrapezial plexus: Both portions of the trapezius obtained their nerve-supply from the subtrapezial plexus. A notable anomaly of this plexus was that it was composed only of the third and fourth cervical nerves; the spinal accessory failed to pierce the sternocleidomastoid muscle, its fibers terminating within that structure. This condition, while occurring independently of Sprengel's deformity, has been noted by other dissectors of this condition, and may have more significance than has been accredited to it.

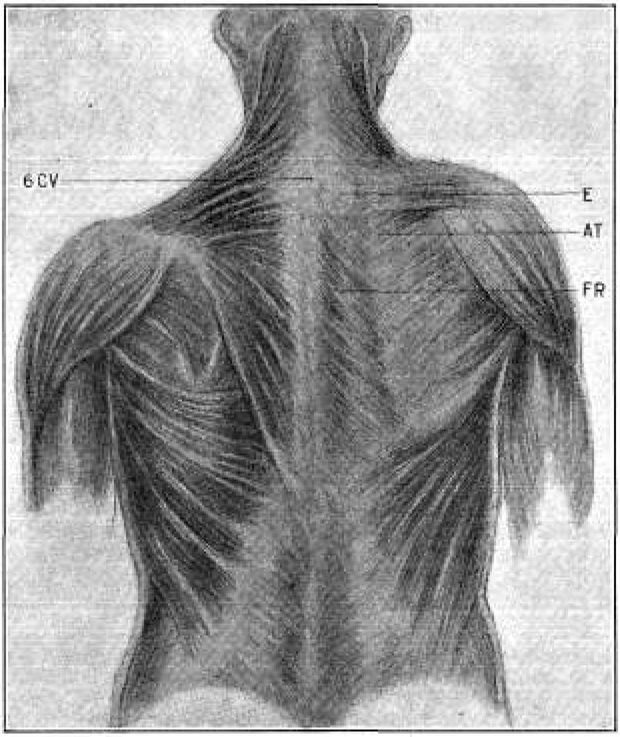

Fig. 4.—Superficial dissection. Shows high position of right shoulder. *6CV*, sixth cervical spinous process; *E*, exostosis; *AT*, anomalous portion of trapezius; *FR*, fibrous tissue replacing thromboid muscles.

The levator anguli scapulæ: This muscle was slightly below normal in size and branched toward its insertion in the form of a Y. One arm of the Y inserted into the superior angle of the scapula and the other into the bony arch. Not over 4 cm. separated the two insertions. If the two arms were spread so as to convert the Y into a T the cross-bar to the T would measure about 5 cm.

The rhomboidei major and minor: These muscles as such were absent, their place being occupied by white connective tissue whose fibers took the same general direction as those of the missing muscles would have done.

The serratus magnus: This muscle was continued upward and inward beyond the vertebral border of the scapula, and interlaced with the rhomboid fibers, making a strong network in which two layers of rather stringy muscle fibers lay.

Upon cutting the scapula away from the trunk two slips from the serrarus magnus were encountered, superior to the main muscle and its attachment to the vertebral border of the scapula. One of these inserted into the superior angle of the scapula, and had its origin from the scalenus anticus, medius, and posticus muscles. The other arose from the first rib and the scalenus posticus and inserted into the vertebral border of the scapula.

Anomalous muscles: Immediately posterior to the levator anguli scapulæ was a small slip of muscle which had its origin from the cervical aponeurosis opposite to the fourth cervical vertebra. Its insertion was very close indeed to that of the levator anguli scapulæ and the fibers ran in the same direction, but the two were not continuous. It was supplied by a twig from the fifth cervical nerve, which sent branches into the levator also, so that a fair conclusion would seem to be that it was an accessory levator anguli scapulæ, having its origin from the cervical aponeurosis.

From this description it will be seen that the elevated right scapula could have been movable only to a very slight extent. Most of the muscular attachments would tend to pull the bone upward or at least help to retain it in its high position, and a glance at the scapula or the bony arch will show that there was great muscular strain on these parts.

SOME REMARKS ON THE TREATMENT OF AMEBIC DYSENTERY.

By George M. Niles, M.D.,

PROFESSOR OF GASTRO-ENTEROLOGY AND CLINICAL MEDICINE, ATLANTA MEDICAL COLLEGE, ATLANTA, GEORGIA.

Within the last two or three years the scientific view-point concerning amebic dysentery and its treatment has undergone a decided change. While there may be present concomitant pathological conditions requiring appropriate attention, it is recognized that the crux of the whole therapeutic problem in this form of dysentery is to destroy the amebæ, and later to heal the ulcerated intestines and cure other complications.